Vascular malformations - therapeutic options

Clemens, Robert

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Disclosure

Speaker name:

Robert K. Clemens

I have the following potential conflicts of interest to report:

- ☐ Consulting
- ☐ Employment in industry
- ☐ Stockholder of a healthcare company
- ☐ Owner of a healthcare company
- ☐ Other(s)

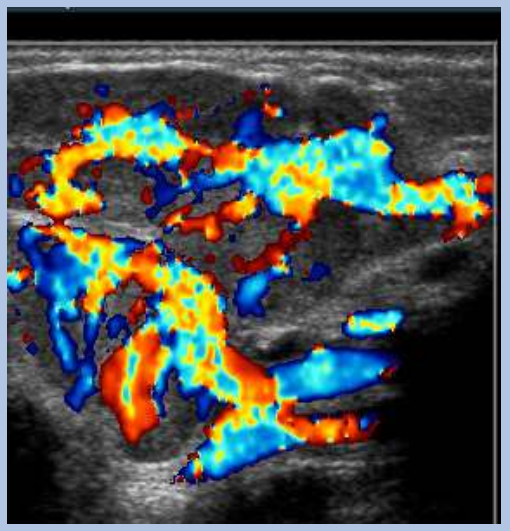
- ☒ I do not have any potential conflict of interest

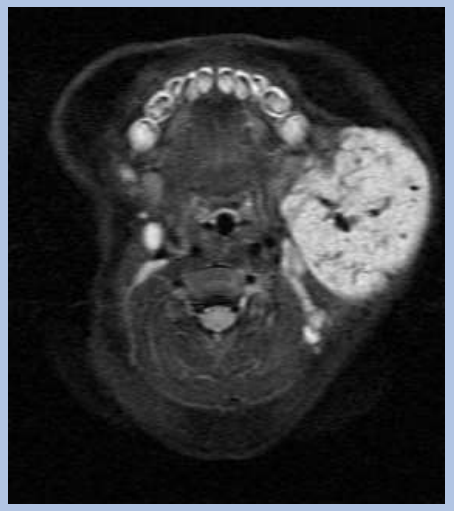
Terminology

- «Cavernous hemangioma» > Venous malformation
- «Cystic hygroma» > Lymphatic malformation
- Term «Hemangioma» misused (71.3%)
- Chance of wrong treatment in 20.3% of cases

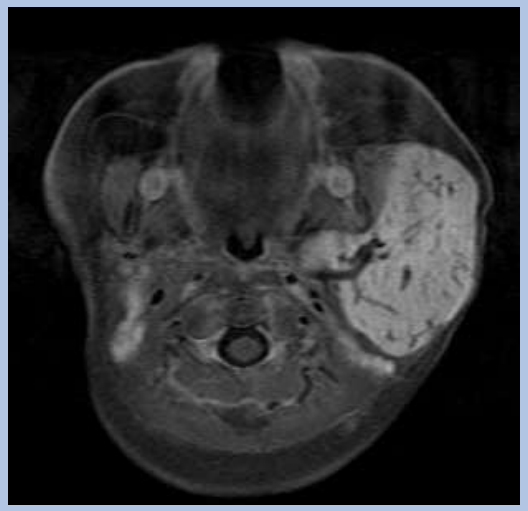
Hassanein AH, Mulliken JB, Fishman SJ et al.Evaluation of terminology for vascular anomalies in current literature.Plast Reconstr Surg. 2011 Jan;127(1):347-51.

Imaging

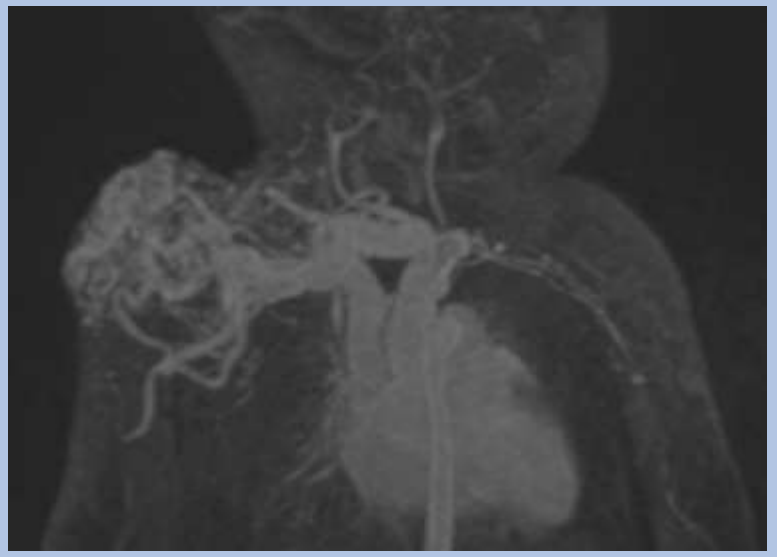


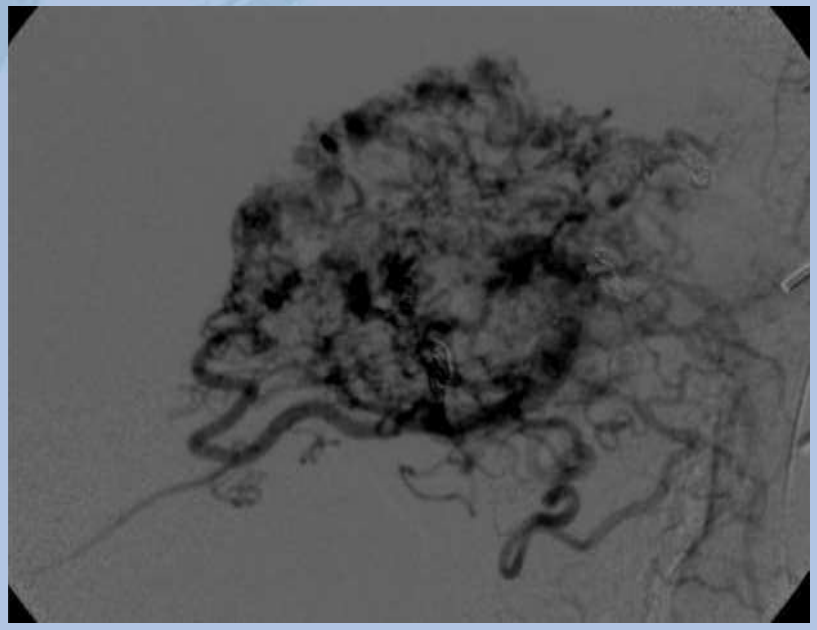


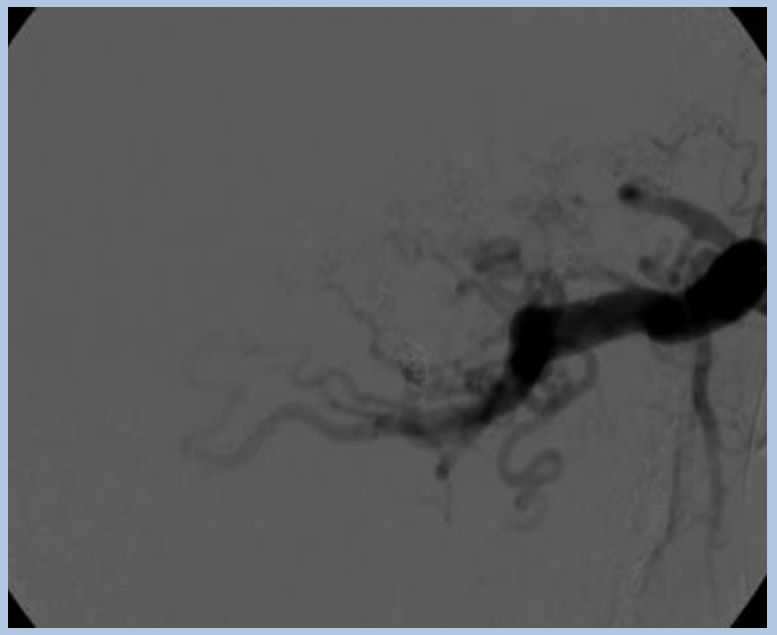
MRI: T2-weighted fat-sat.



T1-weighted image post KM



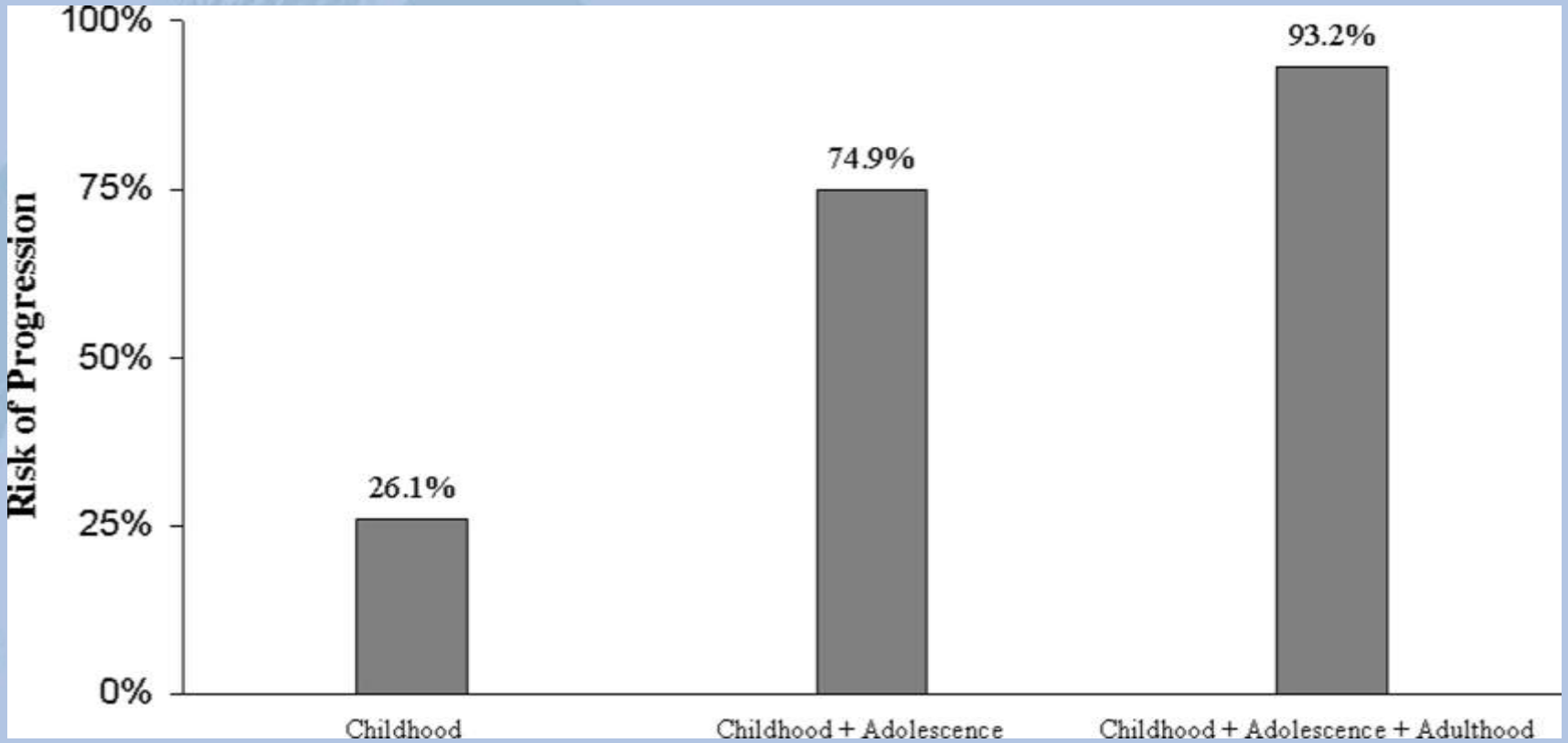




Venous Malformation

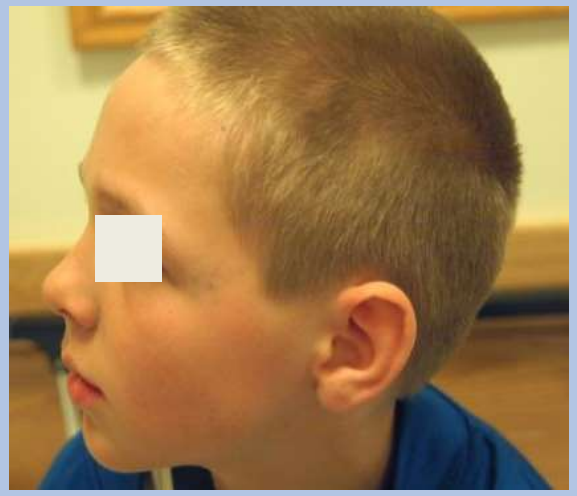
- Most frequent slow flow malformation
- Congenital (often seen at birth)
- 90% with cutaneous involvement with bluish appearance of cutaneous lesions
- Somatic mutation (except GVM and cutaneomucosal venous malformations)

Risk of Progression in VM



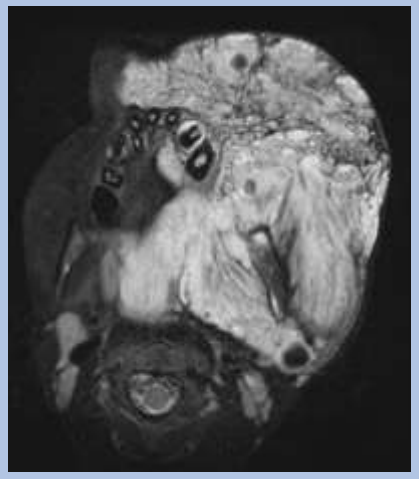
HASSANEIN AH, MULLIKEN JB, FISHMAN SJ ET AL. VENOUS MALFORMATION: RISK OF PROGRESSION DURING CHILDHOOD AND ADOLESCENCE. ANN PLAST SURG. 2012 FEB;68(2):198-201.

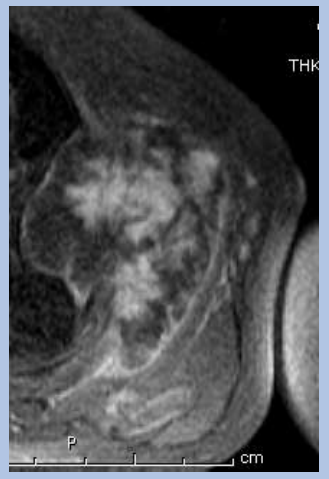
Venous Malformation



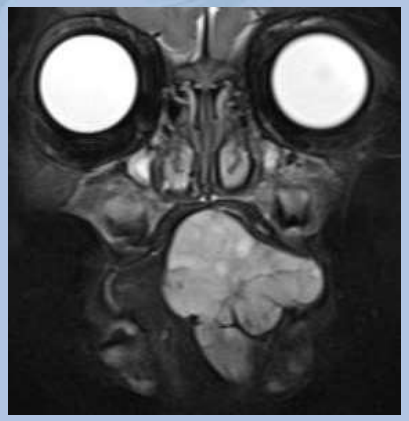
Imaging features

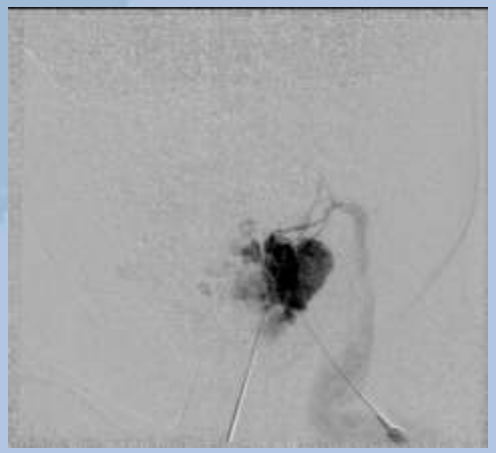
- **MRI:**
  - Phleboliths
  - Thin membranes
  - Fluid filled spaces
  - Fluid-Fluid levels
- **US:**
  - Compressible
  - Slow venous flow, if any

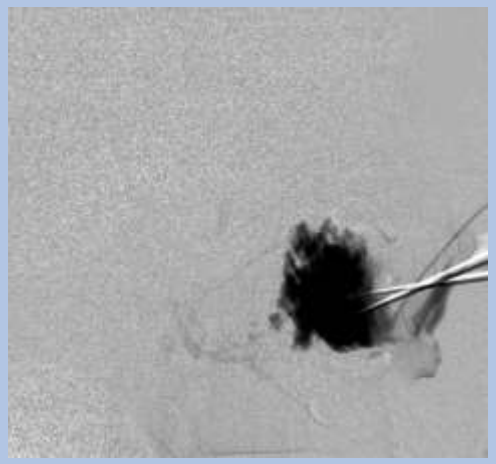




Treatment with Sclerotherapy





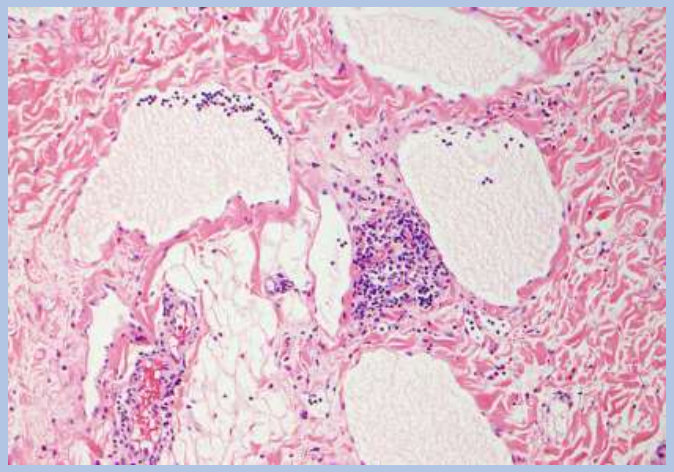


2 YOF with multiple VMs including large tongue lesion.

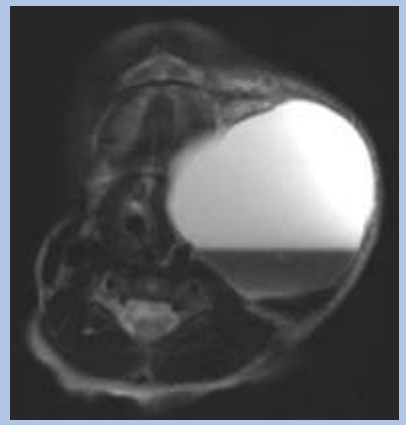
1st procedure: STS 5 mL, ETOH 3 mL

Lymphatic Malformation

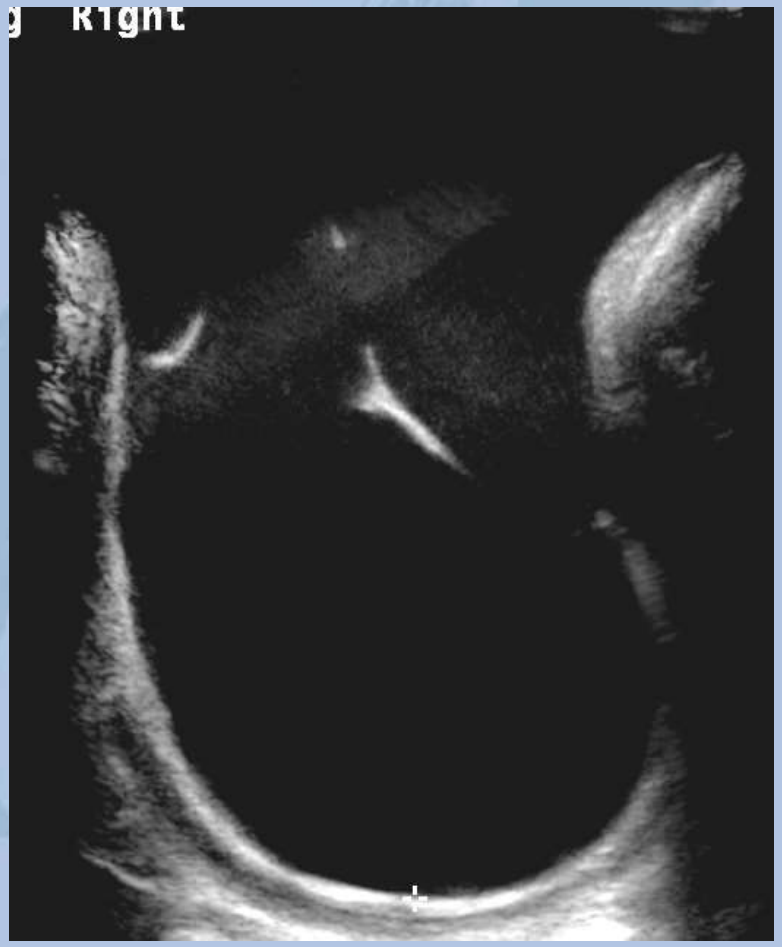
- Abnormal development of the lymphatic vessels
- Common sites: neck, axilla, and pelvis
- Three subtypes:
  - Microcystic, macrocystic, combined
- Intermittent swelling:
  - Intralesional bleeding, Infection, systemic illnesses

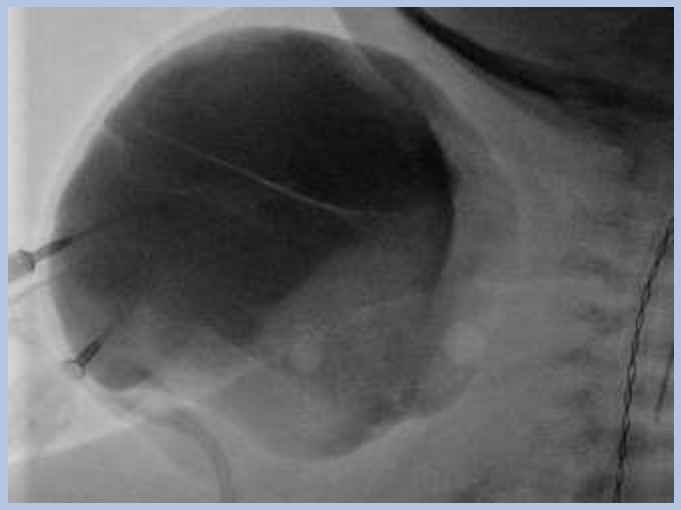


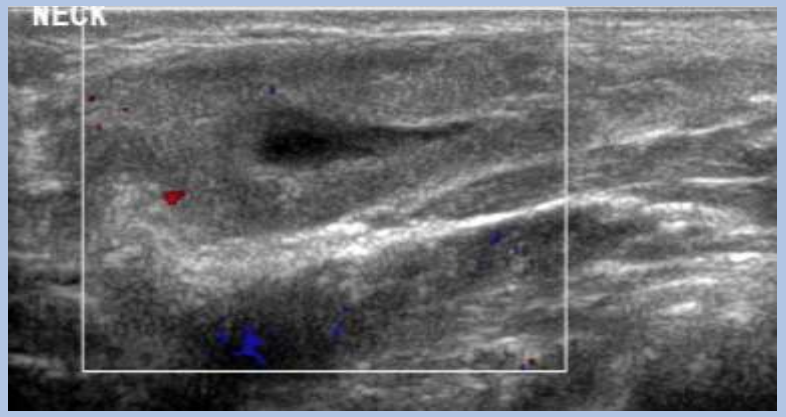
Macrocystic Lymphatic Malformation



Treatment

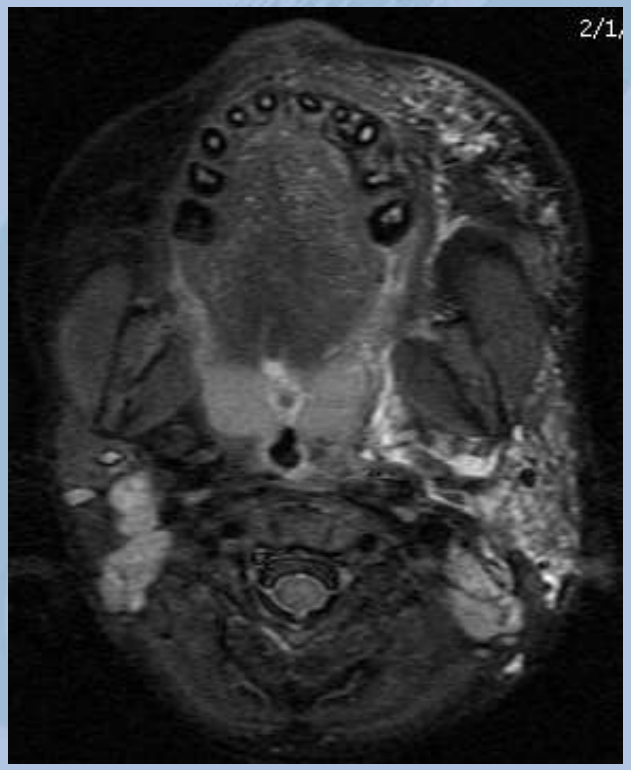


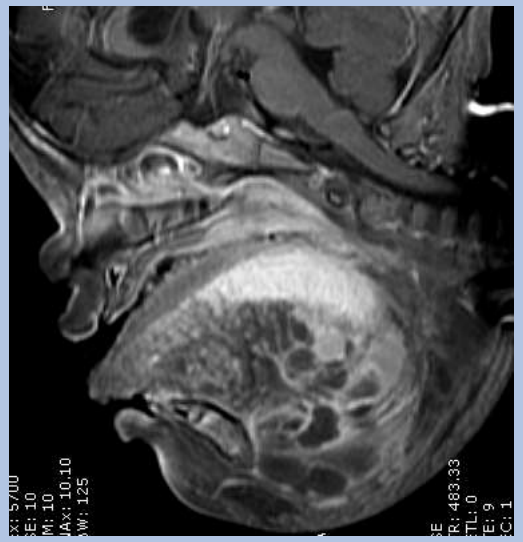




Sclerotherapy: Doxycyline, Ethanol

Microcystic Lymphatic Malformation



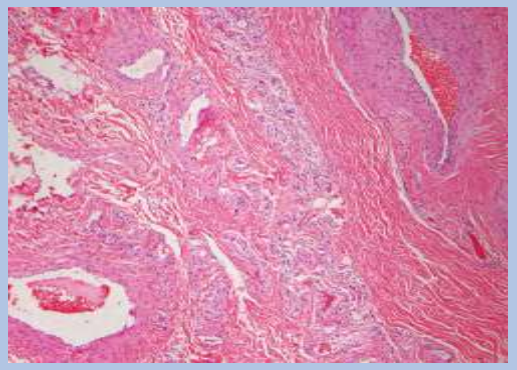


Sclerotherapy: Bleomycin, Doxycyline, Ethanol

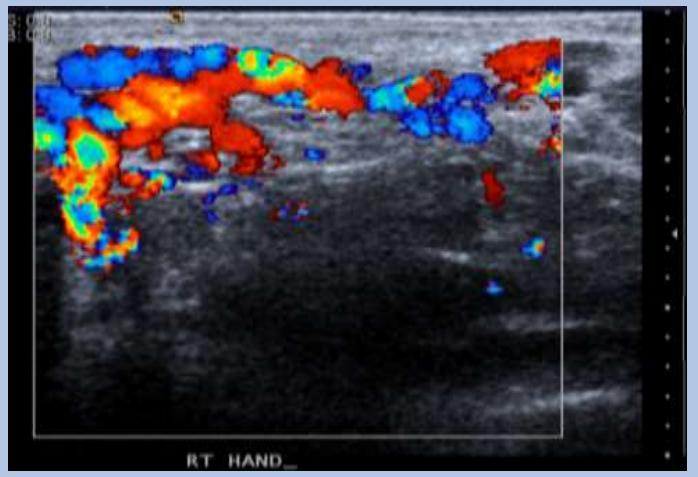
Arteriovenous Malformation (AVM)

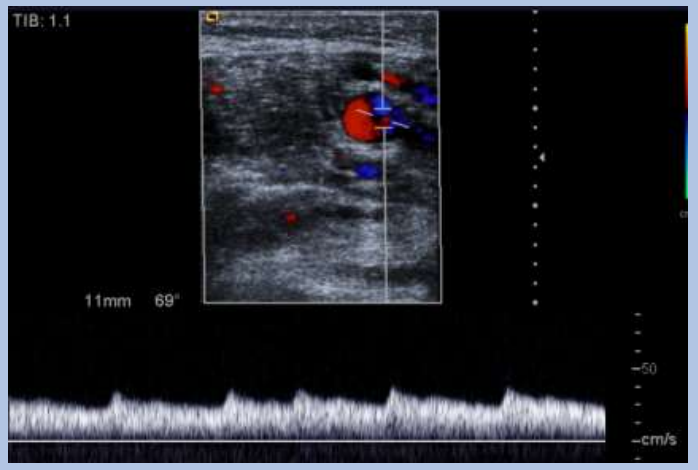
Four clinical stages (Schobinger):

1. Quiescent
2. Expansion
3. Destruction (ulceration, bleeding and pain)
4. High outflow heart failure (rare)



Arteriovenous Malformation (AVM)






i.v. KM

Risk of Recurrence

	Embolization Only (254 Treatments in 102 Patients)		Resection with or without Embolization (118 Treatments in 98 Patients)	
	Probability (%)	95% CI (%)	Probability (%)	95% CI (%)
Overall	98	95–100	81	73–87
Stage I	80	47–94	21	6–52
Stage II	99	95–100	85	73–93
Stage III	99	94–100	81	69–88
Stage IV	100	100–100	—	—

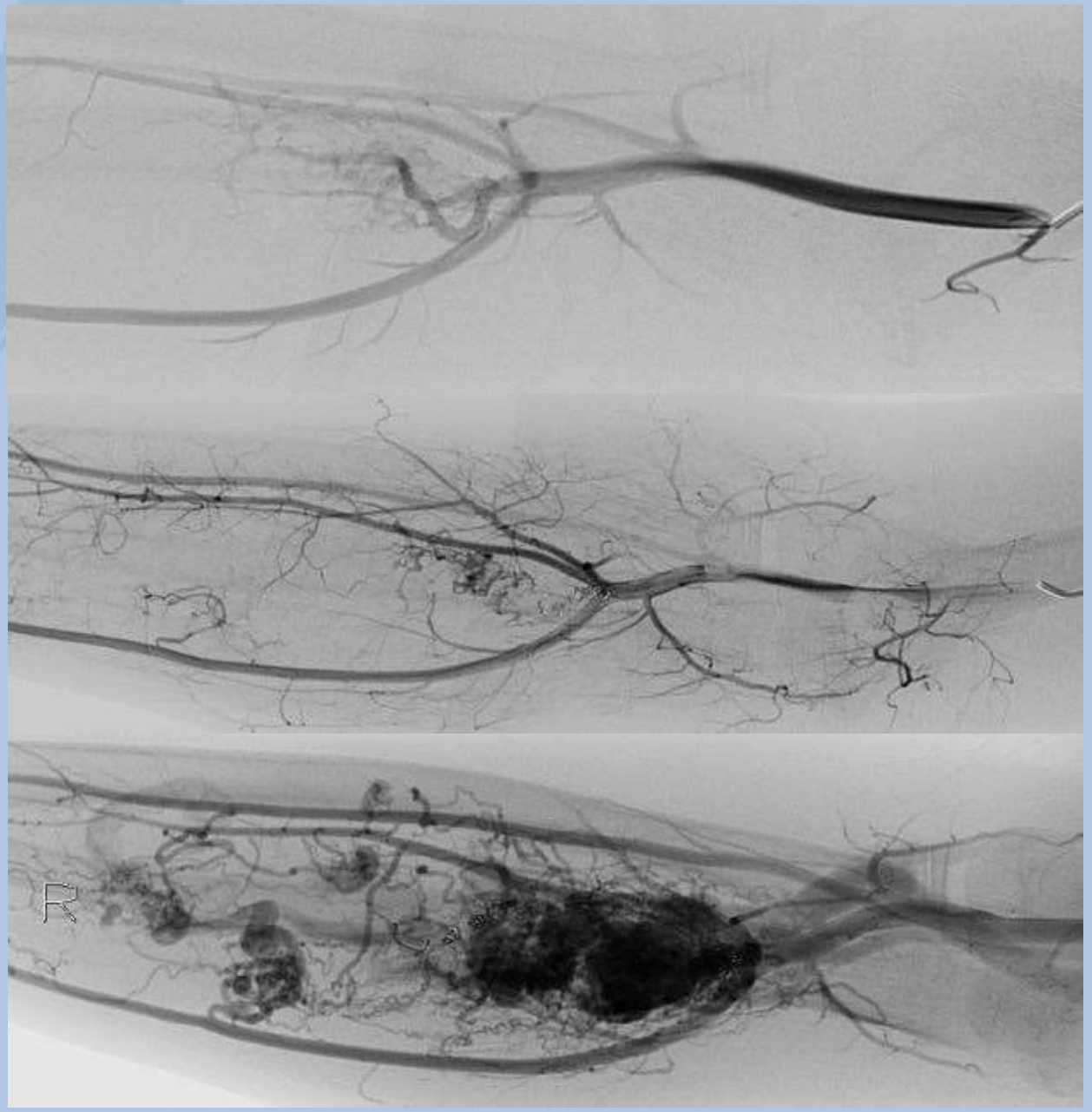
LIU AS, MULLIKEN JB, ZURAKOWSKI D ET AL. EXTRACRANIAL ARTERIOVENOUS MALFORMATIONS: NATURAL PROGRESSION AND RECURRENCE AFTER TREATMENT. PLAST RECONSTR SURG. 2010 APR;125(4):1185-94.

Embolization Nidus (AVM)

Pre embolization

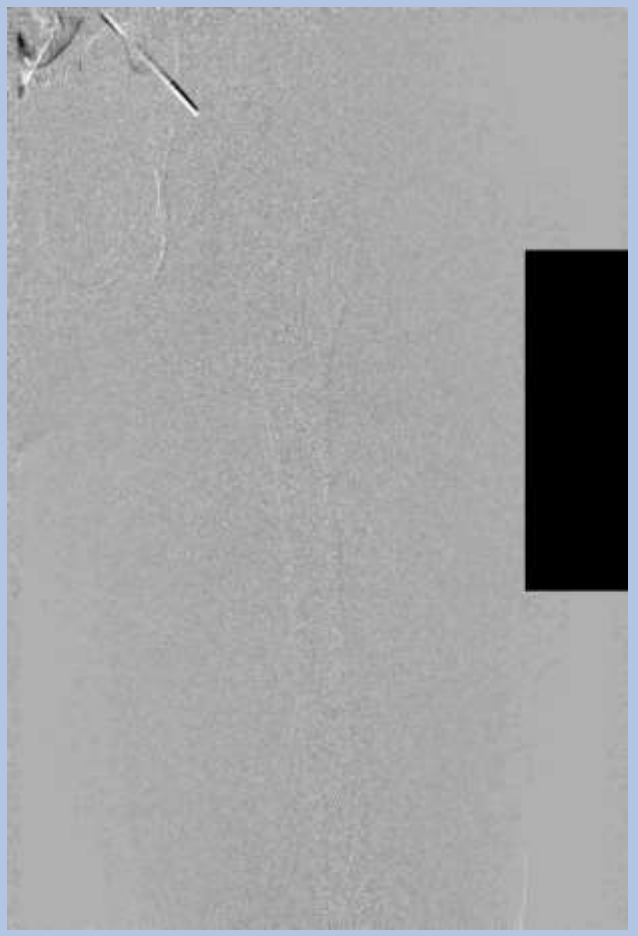
Post embolization

3 months later

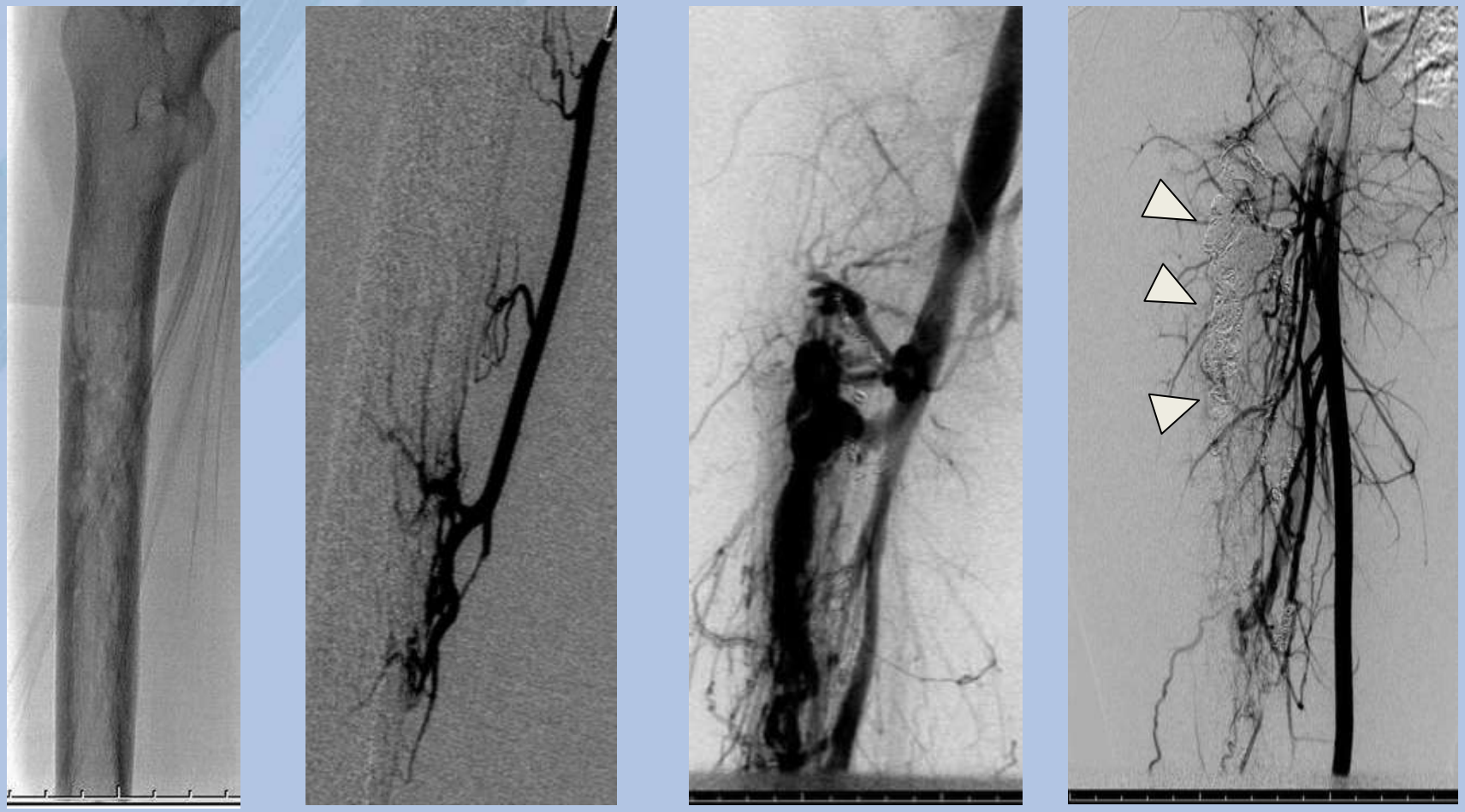


Arteriovenous Malformation (AVM)



Embolization of draining vein (AVM)



Clemens RK, Alomari AI. Transcutaneous treatment of an intraosseous arteriovenous malformation. *CIRSE* 2013. Abstr. Book:394.

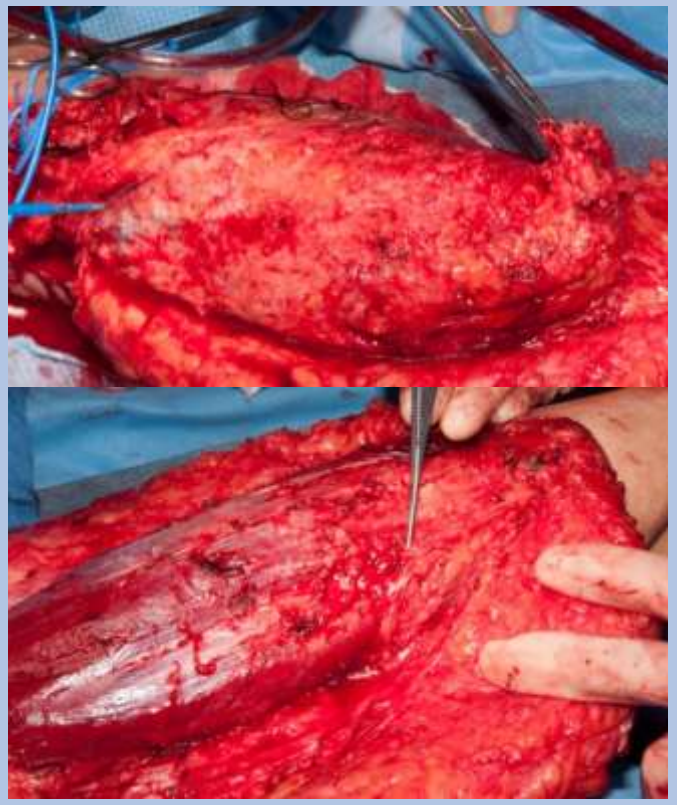
Medical treatment of vascular malformations

- Limited, off-label use
- Addition to surgical and/or interventional treatment
  - Reduce risk of recurrence
  - Enhance primary treatment effect

  - m-TOR-inhibitor (Sirolimus) for AVMs and BRBNS
  - PI3K inhibitors
  - Bevacizumab and interferon reduce venous recanalization

Summary

- Interventional
  - First or additional
  - Embolization for fast-flow
  - Sclerotherapy for slow-flow
- Surgical
  - Total excision possible
  - Overgrowth, tissue
  - «Deal» > function, esthetic
- Medical
  - Limited, additional, off-label use



RARELY CURABLE, CONTROL OF SYMPTOMS, MULTIPLE SESSIONS

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